CHRONIC CONSTRICTIVE PERICARDITIS.

I. Some Clinical and Laboratory Observations

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The cases which are described here in some detail constituted the patient material for an electrokymographic and roentgenkymographic study which is reported in an accompanying paper. Clinical features, angiocardiograms, cardiac catheterization data, renal clearance determinations, and results of pericardectomy are of considerable interest and are presented in this report.

Case 1. S. W. A. In 1923 this white male patient, born in 1897, had, for several months, chest pains so severe he "had to take 18 aspirins a day". A positive serologic test for syphilis was found and treatment instituted. He was well until 1945 when there was onset of symptoms consisting of fever, chills, weakness, and progressive abdominal swelling. He was a confirmed alcoholic. For two years he was believed to have Laennec's cirrhosis. Several "peritoneal button" operations were performed with limited benefit. At the time he was studied in 1948 shortness of breath, ascites and leg edema were the main manifestations. He had been unable to work since the onset of his illness three years previously. The main features of the physical examination were limited thoracic expansion, greatly distended veins of the neck and arms, liver enlargement to a point 5 inches below the right costal margin, and slight cardiac enlargement. The blood pressure was consistently about 110/80 mm. Hg and there was a slight but definite pulsus paradoxus. X-rays revealed a dilated superior vena cava, slight cardiac enlargement and no pericardial calcification. The arm venous pressure was 320 mm. of saline with no significant change after digitalization. The arm-to-tongue circulation time (Decholin) was 18 seconds. Operation was performed on November 4, 1948, by Dr. Alfred Blalock. Through a median sternotomy incision the entire anterior surface of the heart was exposed. There was dense fibrosis in the groove between the right ventricle and right auricle and this fibrous process encircled the right auricle and both venae cavae in an extensive manner. There was almost no involvement of the ventricles themselves. Over the right ventricle the pericardium was thickened but there was no evidence of involvement of the epicardium in this area. The pericardium bound to the right auricle and venae cavae and a large portion of the pericardium overlying the right and left ventricles were successfully removed. Post-operatively the venous pressure fell to normal in the first week, the liver shrank to a position just below the costal margin, and there was no further evidence of fluid accumulation. Five months following operation the patient returned to a full work program. He has remained well almost three years following operation and works as a merchant seaman.

Case 2. R. McA. This is a white male patient born in 1922. In 1942 the patient developed transient episodes of nausea, vomiting, weakness and easy fatigability. These episodes continued for about two years. In 1945 he was rejected by the service because of "organic heart disease". The patient was able to do unusually heavy labor without difficulty until 1948

when after a period of exertion he became nauseated, "weak in the knees", and lost consciousness. His physician told him his heart "beat a little too fast" and prescribed digitalis. In the next few months the patient began to feel progressively less well and there was weight gain with abdominal swelling. Significant physical findings preoperatively in 1949 were moderate generalized venous distension (venous pressure 270 mm. saline), enlargement of the heart to the left and right, auricular fibrillation, split and accentuated second pulmonic sound, split first sound at the apex, liver down three cm., bilateral varicoceles, and blood pressure 100/70. Digitalization had no effect on the elevated venous pressure. X-rays revealed very extensive calcification of the pericardium. On February 16, 1949, pericardectomy was performed by Dr. Alfred Blalock using a median sternotomy incision. To the left the decortication was extended far around onto the left ventricle and on the right as far as the venae cavae; inferiorly the diaphragmatic surface was freed as far as the central tendon. Only a plaque of calcium in the region of the left anterior descending artery was left. When restudied in April 1949, venous pressure was 75 mm. in the right arm, 135 mm. in the left; the liver was down 2 cm. and there was no fluid accumulation. In June 1950 auricular fibrillation had reappeared. However, the liver was not palpable, heart sounds were normal, and there had been no edema or unusual dyspnea. The patient had been doing heavy labor. Venous pressure was 80 mm. saline. In May, 1951, the findings were the same. He had been maintained on digitalis during the previous year and had not missed a single day from his work with a construction crew.

Case 3. A. N. This is a white male patient born in 1897. His pertinent history begins in 1920 at which time he developed shortness of breath and abdominal swelling, was hospitalized for 9 months and told he had "water on the heart". He recovered from that illness, however, without significant physical disability. In 1936 "overnight" he began to have abdominal swelling, leg edema and dyspnea. In 1944 a partial pericardectomy was performed at a New Orleans hospital using a left precordial incision. The decortication was considerably limited by serious bleeding, probably due to a small rupture of the left ventricle. Thereafter, however, he had almost complete relief of symptoms and no longer required frequent abdominal paracenteses as before. In 1946 again "overnight" his previous symptomatology returned. With this he complained of severe precordial pain for which opiates were given to the point of addiction. These were his complaints when studied in 1949. The positive physical findings were greatly distended neck and arm veins, limited chest expansion, enlargement of the heart to a point 2 cm. beyond the left midclavicular line, blood pressure 108/80 with slight but definite pulsus paradoxus, a protodiastolic sound, liver down 5 finger-breadths. In the left mid-precordium at the site of the rib defect, the cardiac impulse was quite forceful. There was an outward movement with ventricular filling which was quick and slapping, making it possible to suspect the "flat-top and V's" EKY pattern described elsewhere (2). X-ray of the chest revealed extensive pericardial calcification, particularly on the diaphragmatic surface, cardiac enlargement, a dilated superior vena cava, and extensive pleural thickening bilaterally. Fluoroscopy revealed reduced pulsations everywhere except for an area at the cardiac apex which appeared almost herniated and which had a gross "flat-top and V" type of movement. Venous pressure was 275 mm. saline, vital capacity (3 sec.) 1.0 liter, arm-to-tongue circulation time 40 sec., arm-to-lung circulation time 26 sec. On September 7, 1949 pericardectomy was performed by Dr. J. L. Southworth via a median sternotomy. All portions of the heart visualized except for the apex itself were involved in a very dense calcific and fibrotic process. The entire anterior portion of the right ventricle, right auricle and venae cavae was decorticated. Post-operatively the liver receded moderately in size, venous pressure fell to 130 mm. saline, arterial pressure rose to 130/70 and the previously rapid fluid accumulation ceased. Five weeks following operation auricular

flutter appeared and the day following this event the patient developed a right hemiplegia which was believed to be due to embolism from the left auricle. Recovery from this accident was very slow with respect to both speech and the use of arm and leg. It did have one desirable effect: the patient's addiction disappeared completely.

Case 4. L. P. W. In October 1949, this colored male patient born in 1902 had the onset of an acute febrile illness characterized by productive cough and bilateral pleuritic pain. He was studied at a Washington, D. C., hospital and found to have a left pleural effusion and a large pericardial effusion. These slowly cleared and he was able to return to work in February 1950. At about this time, however, he noted swelling of the face in the morning and of the ankles at night. A prominent protodiastolic third heart sound was recorded. Abdominal swelling and exertional dyspnea appeared and he found it necessary to sleep on two pillows. In July 1950, significant findings included B. P. 135/110 with pulsus paradoxus, generalized venous distension, right pleural effusion and ascites, hepatomegaly and hyperbilirubinemia. On July 19, 1950, median sternotomy was performed by Dr. Richard Kieffer who found a dense fibrous scar encasing the entire heart. The heart was rotated clockwise and mainly the right ventricle and a portion of the right auricle were decorticated. The posterior part of the heart—mainly left ventricle—was freed from the overlying pericardium by blunt dissection. Ventricular fibrillation developed during operation but was successfully reverted with cardiac compression and procaine amide (Pronestyl Squibb). Histologically the pericardial scar showed tubercles. Streptomycin, 1 gm. a day, was started five days before operation and continued for a total of two months. Following a stormy post-operative course the patient gradually improved. Venous pressure returned to normal and the liver receded to the costal margin. After nine months of rest he returned to work as a teacher.

Case 5. C. L. M. This white male patient born in 1891 had no history of acute pericarditis or tuberculous disease. There had been rheumatoid arthritis of moderate severity for ten years beginning in 1940. In August 1949, the patient sustained a fracture of two vertebrae as a result of a steam-boiler explosion which threw him several yards. He was rendered unconscious for two hours but recalls no symptoms referable to the heart or lungs. Later in 1949 there was the onset of occasional evening pedal edema. In June 1950, there was the abrupt onset of severe right upper quadrant pain and sensation of fullness of the head when he leaned over or lay flat. The most significant findings in July 1950 were generalized venous distension (venous pressure 270 mm. saline in arms and legs), slight cardiac enlargement, scratchy heart sounds with a protodiastolic sound, hepatomegaly, and a dilated superior vena cava by x-ray. Pericardectomy was performed on July 28, 1950 by Dr. Alfred Blalock using an incision in the fourth left interspace. Overlying the right ventricle and right auricle and between these chambers and the sternum was a large, thick-walled cyst containing 300 cc. of material which resembled dark coffee. The walls of the cyst were parietal and visceral pericardium; the cyst resembled a subdural hygroma. In other areas the pericardium was adherent to the epicardium and was thickened. A wide decortication of both ventricles was performed. Following operation venous pressure fell promptly to normal, the liver decreased in size, fluid accumulation ceased, and all symptoms—particularly the sensation of fullness in the head—disappeared. Post-operatively, coincident with the decrease in liver size, a conspicuous hepatic friction rub was observed over a two week period. The patient has now remained well one year following pericardectomy.

Case 6. L. N. N. This is a white male patient born in 1934. An older brother has had pulmonary tuberculosis. At the age of two or three years the patient had cervical adenitis and fever for several weeks. At the age of 13 he developed a severe productive cough and fever persisting for 10 days. Shortly thereafter a pulmonary lesion was found and interpreted as tuberculosis, although no organisms were identified and the tuberculin skin test was said

to be negative. In 1949 he developed anorexia, nausea, vomiting, easy fatigability, headache and fever. This persisted for one month, gradually subsiding. Three months later abdominal swelling was noted. This progressed to the point that paracenteses were necessary. Ankle edema appeared. When first seen in December 1949, examination revealed low grade fever, tachycardia, generalized venous distension, hepatomegaly, ascites, and ankle edema. Significant laboratory data included alkaline phosphatase 11.5 units per hundred cc., bilirubin 3.6 mgms. per hundred cc., 26% BSP retention in 30 minutes, venous pressure of 260 mm. saline in arms and legs, EKG compatible with chronic pericarditis, positive tuberculin skin test (1:1000 O.T.). Because of persistent fever, pericardectomy was deferred. On a second admission in April, 1950, the blood pressure was 90/60 with definite pulsus paradoxus. The right leaf of the diaphragm was elevated. The sounds were tic-tac in quality and distant. There was a scratchy systolic sound at the apex varying with respiration. Because of continuing fever and the development of a pleural friction rub operation was again considered inadvisable.

On the third admission in July, 1950, it was found that the patient's condition had deteriorated further with rapid accumulation of fluid and depletion of serum proteins to 4.1 grams per hundred cc. Pericardectomy was performed on July 25, 1950 by Dr. Alfred Blalock using an incision in the fourth left interspace. A dense pericardial scar was successfully removed from the largest portion of both ventricles and a part of both auricles. Grossly the left dome of the diaphragm was studded with tubercles and histologically the pericardial scar revealed tubercles. The patient improved with amazing rapidity. Serum proteins rose from 4.1 to 6.1 (albumin from 2.3 to 3.5 gms. per hundred cc.) in two weeks. Venous pressure fell to normal in the first twenty-four hours and remained so. Fluid accumulation ceased. The patient was treated with dihydrostreptomycin and para-amino-salicylic acid for 50 days. The patient is considered a clinical cure.

On Thanksgiving Day, 1950, the patient sustained trauma to the head. This was followed by development of manifestations of tuberculous meningitis for which he has been treated with streptomycin intramuscularly and intrathecally, together with para-amino-salicylic acid intravenously. His course has been satisfactory and it is anticipated that he will recover. This experience indicates the necessity for prolonged anti-tuberculosis therapy in cases such as this with active disease at the time of operation (1).

Case 7. W. B. This white boy, born in 1934, had very acute development of abdominal swelling with no other symptoms one month before first being seen in August, 1950. He had previously been in excellent health and engaged actively in sports particularly baseball. He was exposed to the maternal grandfather and a maternal uncle, both of whom died of tuberculosis. Examination revealed generalized venous distension (venous pressure 330 mm. saline), mulberry cyanosis of hands, feet, lips and ears, blood pressure 110/90 with definite pulsus paradoxus, right pleural effusion and ascites, a calcified placque over the left ventricle, enlarged left auricle and dilated superior vena cava by x-ray, liver down four finger-breadths. In the left mid-precordium there was a visible and palpable impulse with suggestively "flat-top and V" characteristics (2). There was a conspicuous protodiastolic heart sound. Pericardectomy was performed by Dr. Henry Bahnson on September 11, 1950, through an incision in the fourth left interspace. The exposure permitted resection of scar from the venae cavae anteriorly to a point well around posteriorly on the left ventricle. There was no clue as to the etiology of the process. Venous pressure fell to about 140 mm. water following operation, fluid accumulation ceased, and the patient has been asymptomatic for ten months.

Case 8. J. M. This was a colored male patient, born in 1893, who in 1944 had onset of leg edema and dyspnea. This increased slowly but progressively until the time of his admission to the hospital in 1949. Significant physical findings were marked generalized venous

distension, enlargement of the heart to right and left, auricular fibrillation, a superficial crunchy pericardial sound in the left mid-precordium, liver 10 cm. below the right costal margin, bilateral varicoceles, ascites, and 2 plus ankle edema. The blood pressure was 104/90 mm. Hg, the venous pressure 370 mm. saline. X-rays revealed extensive pericardial calcification. The cardiac apex was free of calcification and on fluoroscopy showed a distinct "flat-top and V" type of movement (2) of quite large amplitude. EKG's showed auricular fibrillation and right axis deviation. On January 16, 1950, exploration was performed by Dr. J. L. Southworth via a median sternotomy incision. There was a very extensive calcified shell surrounding all portions of the heart visualized. Decortication was undertaken and when approximately one-half the anterior cardiac surface had been freed the right ventricle which had ballooned up during the procedure ruptured. Attempts to repair the large rent only made it larger, the myocardium being too friable to hold sutures. The patient exsanguinated. Autopsy confirmed the clinical impressions: the left auricle was considerably dilated. There was no definite evidence of tuberculosis.

Case 9. N. D. This white male patient born in 1915 had a sister with pulmonary tuberculosis treated by pneumothorax. At the age of fourteen, there was an illness characterized by sudden prostration, chest pain, and cough following which dyspnea on exertion was noted. At the age of 16 a tentative diagnosis of tuberculosis was made and sanatorium care recommended. At age 18 dyspnea became worse; ankle edema was noted and he was told he had a weak heart. Dyspnea on unusual exertion continued and became gradually worse. Physical examination in May, 1949, revealed moderate venous distension in the neck, heart of normal size, liver down five finger-breadths. Laboratory data included venous pressure varying between 138 and 235 mm. saline, circulation time 23 seconds (Decholin), and a triangular, slightly enlarged heart with calcification of the postero-inferior portion of the pericardium. The general physical condition was the same or slightly improved when studied in detail again in November, 1949, and in February, 1950. On examination in June, 1950, it was found that the patient's only symptom was dyspnea on unusual exertion. The liver was down two finger-breadths but there was no ankle edema. X-rays revealed a dilated superior vena cava, enlargement of the heart to the right, and pleural thickening on the right. Venous pressure was 160 mm. saline. (Operation was decided against because of the patient's relatively slight incapacitation.) His status was identically the same in July, 1951.

Case 10. W. M. This white male patient born in 1893 had onset of easy fatigability late in 1948 and blueness of the lips and nails late in 1949. He was told by a physician that there was calcium around his heart. There was occasional slight ankle edema. He was digitalized without evident benefit. There was one syncopal attack of five minutes duration following descent of a flight of stairs. The patient's health had been excellent prior to the development of the above symptoms. There was no known exposure to tuberculosis. Physical findings included blood pressure 180/100, cyanosis of lips, ears and nailbeds, distention of the neck veins, a prominent protodiastolic sound, liver down three finger-breadths. Laboratory findings included venous pressure 140 mm. saline, normal hemogram (Hgb. 15 gms.), arterial oxygen saturation 87%, heart normal in size by x-ray with extensive calcification overlying both ventricles and 10% methemoglobinemia (the last subsequently disappeared). In an attempt to improve the patient's functional status pericardectomy was attempted on September 18, 1950, by Dr. Alfred Blalock using an incision in the fourth left interspace. From the beginning heart action was seen to be very feeble and cardiac standstill occurred before any decortication could be performed. Autopsy revealed a very extensive *ossified* shell about both ventricles (*Panzerherz*) with accentuation in the atrio-ventricular grooves. Both auricles were free and considerably dilated. The myocardium of both ventricles was very atrophic. No definite evidence as to etiology was discovered.

Case 11. N. F. This patient was a rabbi born in Russia in 1876. Because of language dif-

ficulties, it was impossible to determine whether any illness suggestive of acute pericarditis had occurred in previous years. He was well until 1947 when there was onset of dyspnea and ankle edema. He was treated with digitalis with benefit. Examination in June, 1950, revealed distended neck veins, blood pressure 160/96, auricular fibrillation, a harsh systolic murmur at the apex, liver margin at the level of the umbilicus, 3 plus pitting ankle edema. X-ray of the chest revealed a dilated superior vena cava, right pleural effusion, and pericardial calcification in the inferior and left borders of the heart. Venous pressure was 280 mm. saline and never was less than 200 mm. when fully digitalized and after mercurial diuresis. Surgery was considered to be inadvisable because of the patient's age.

The patient was admitted to the hospital in August, 1950 for treatment of his moderately refractory edema. He died unexpectedly in his sleep a few days after admission.

Case 12. M. T. This colored female patient born in 1899 had spring and summer asthma from 1939 to 1945. Between 1943 and 1948 she was followed in a city clinic because of a lesion in the apex of the left lung which was believed to be quiescent. Several members of the family had died of tuberculosis. In June, 1948, there was onset of weakness, easy fatigability, heavy feeling in the abdomen, attacks of coughing with vomiting, and ankle edema. Examination in December, 1948, revealed blood pressure 110/80, palpable apical impulse and liver to the level of the umbilicus. Observations of venous pressure varied between 160 and 250 mm. saline. The heart was normal in size with some prominence of the pulmonary artery. Because of continued low-grade fever and tachycardia the patient was transferred to a sanatorium where she remained for six months. In June, 1950, it was discovered that the patient had felt quite well since discharge from the sanatorium in September, 1949. She had, however, continued to have exertional dyspnea and ankle edema as well as "bloating of the stomach". There was distention of the neck veins, an accentuated P2 and a scratchy systolic sound over the left mid-precordium. The liver extended 3 cm. below the right costal margin. X-ray revealed no pericardial calcification. The heart was normal in size with prominence of the pulmonary artery. Venous pressure was 280 mm. saline. Operation was postponed because of the patient's age and relatively insignificant symptoms.

Case 13. J. H. This white male patient born in 1925, first had manifestations of constrictive pericarditis in 1941 when enlarged neck veins were noted. His original admission for pneumonia of the right lower lobe due to type 2 pneumococcus was in November, 1946. The classic signs of constrictive pericarditis were discovered. Jaundice accompanied the pneumonia. There was an overactive precordium at the apex where a palpable diastolic impulse was described. The blood pressure was 90/70 with a distinct pulsus paradoxus. The liver was down 2 finger-breadths. There were bilateral varicoceles. There was 2 plus ankle edema and questionable ascites. Venous pressure before the first operation was 360 mm. saline and the circulation time (Decholin) 25 seconds. Total proteins and A/G ratio were normal. At one time the prothrombin time was 52 seconds (less than 25% of normal). BSP retention in 30 minutes was 44% and the serum cholesterol was 83. EKG revealed T2 and 3 inverted, T4 biphasic. X-ray of the chest revealed the heart to be at the upper limit of normal for size. There was linear calcification in the left border. On January 11, 1947, the first pericardectomy was performed by Dr. Alfred Blalock. The incision was made to the right of the sternum with removal of several costal cartilages. A greatly calcified pericardium was encountered. The right auricle and right ventricle were decorticated. One week following the procedure the venous pressure was 170 mm. saline, and the liver had diminished somewhat in size. However, the venous pressure remained between 170 and 250 mm. saline. Therefore, on July 28, 1947, a second operation was performed. On the left, costal cartilages 3 to 6 were removed and the left and anterior surfaces of the heart were freed of a heavy calcium deposit. Even after this second operation there occurred no definite improvement in either

venous pressure or the tendency to accumulate fluid. In the three years since the last operation the edema and ascites have become slowly but progressively more pronounced and refractory to treatment. The patient has complained continually of cold hands and feet, and there is at times obvious cyanosis of the nailbeds and lips. The venous pressure remains over 200 mm. saline at all times. There is no evidence of a specific etiology in this case, although in August, 1949, he developed a chronically draining sinus in the left precordial incisional scar. No acid fast bacilli have been identified in the draining material. Exploration of the sinus tract in July, 1950, suggested that it might be kept open by a sequestrum of calcified pericardium. The sinuses continued to drain and the patient had persistent fluid accumulation requiring mercurial diuretics at least twice weekly. For this reason a third pericardectomy was performed on September 14, 1950, by Drs. Blalock and Bahnson, using an anterior incision in the fourth interspace on the left. Considerable reformation of pericardial scar was discovered. The right auricle, right ventricle, and left ventricle were decorticated in a very extensive fashion. Postoperatively the venous pressure remained elevated, the sinus tracts re-opened, and the manifestations of chronic congestive failure continued unabated.

The patient was re-studied extensively in August, 1951. Normal venous pressure, normal liver size, and absence of edema led us to believe the patient may at last have attained a clinical cure. He is seeking employment for the first time in many years.

Case 14. G. V. S. This is a white female patient born in 1929. At age 5 there occurred a bout of tonsillitis, accompanied by "smothering spells" causing the child to sit up at night. A physician stated at that time that the liver was enlarged. At age 11 ascites developed together with aching epigastric pain and nausea about once a week. At age 12 face edema in the morning was noted as well as epigastric pain on exertion. "Smothering" with exertion had its onset at age 14. Physical examination revealed full fundal veins, absent precordial impulse, a diastolic sound (described as a presystolic murmur by some), an accentuated second pulmonic sound, blood pressure 108/64, questionable pulsus paradoxus, liver down 4 finger-breadths, spleen down 1 finger-breadth. EKG revealed flat T-waves in all leads. X-ray revealed a moderate amount of pericardial calcification. On August 3, 1943, the first operation was performed by Dr. Alfred Blalock. The ends of ribs 4, 5 and 6 on the left and the corresponding costal cartilages were removed. Grossly the heart was bound in a constricting process which appeared to be caseous in places. Because of the "caseous" material, particularly on the right, no attempt was made to expose the venae cavae. Histologically, hyaline scarring and focal calcification were described, but no caseation. The venous pressure was not reduced following operation, although systolic blood pressure did rise to 125 mm. Hg. Exercise tolerance was moderately limited and mercurial diuretics were required for control of fluid accumulation. Because of failure to attain more improvement a second pericardectomy was performed by Dr. Alfred Blalock on August 6, 1947, through a median sternotomy incision. A very extensive decortication was performed freeing especially the right side of the heart and venae cavae. Following this operation, too, the venous pressure did not drop significantly. In November, 1948, the patient gave a history of excellent exercise tolerance. However, the liver was down 4 finger-breadths and the venous pressure was 210 mm. saline; no peripheral edema had been noted. When restudied in May, 1950, the patient was found to be relatively asymptomatic except for RUQ pain and slight ankle edema occurring with upper respiratory infections and just prior to menstruation. Moderate acrocyanosis was observed during an upper respiratory infection. Venous pressure was 200 mm. and she was negative to skin test with O.T. 1:100 (P.P.D., second strength, was negative in 1943). The improved functional capacity was maintained until March, 1951, when ascites and edema of the legs reappeared coincident with a bout of bronchopneumonia. Diuresis was accomplished only

with some difficulty. The venous pressure was consistently over 210 mm. saline. A third pericardectomy was performed on May 1, 1951, through a left anterolateral chest incision in the fourth interspace. Dense scar tissue with calcification encased the heart. Most of the calcified tissue lay posteriorly. A very liberal decortication was performed. Three weeks later the venous pressure was 125 mm. It is believed that she has been much improved by operation, although it is still too early to be certain about long-term results. In October, 1951, the patient was well and active.

Case 15. P. H. This is a white female patient born in 1920. In June, 1947, while she was working in the fields, pain began in the right upper quadrant and radiated downward. Appendectomy was performed. In January, 1948, the right upper quadrant pain became worse and was accompanied by nausea and vomiting. Laparotomy at that time revealed an enlarged liver which, however, was histologically normal. In April, 1948, the principal physical signs were high right diaphragm, greatly distended neck and arm veins, absent precordial impulse, distant heart sounds, liver reaching to the iliac crest, minimal or no ascites and no leg edema. Venous pressure was 275 mm. saline. The only abnormality of liver function was a prothrombin time of 29 seconds. On x-ray the heart was but little enlarged. The superior vena cava was impressively dilated. Operation through a median sternotomy was performed on May 10, 1948, by Dr. Alfred Blalock. A dense fibrous process, one centimeter thick in places, involved the right auricle and right ventricle. A lappet of dense adherent pericardium extended from the diaphragmatic surface upward about one-half inch onto that portion of the left ventricle which could be visualized. The pericardium was densely adherent over the left anterior descending coronary artery and could not be resected at that point. Immediately following operation the venous pressure was 135 mm. saline and it fell quickly to normal thereafter. In November, 1948, venous pressure was 100, the liver was down only 1 finger-breadth and the patient was asymptomatic.

Case 16. M. C. This is a white male patient born in 1928. In December, 1944, he suffered a grippe-like illness characterized by chills and joint pains. In May, 1945, there was onset of swelling of the ankles and abdomen. The patient complained bitterly of coldness, cyanosis, and pain of the feet on relatively slight exposure to cold. Small black areas appeared on the feet, and he was admitted to another hospital for "frost-bite". There was story of fainting following exertion in April, 1943, at which time a physician told him he had low blood pressure. When studied in 1945 the important findings were blood pressure 100/74, distended neck veins with prominent pulsations, a Wenckebach sign (35), distant heart sounds, left pleural effusion, ascites, liver down 4 finger-breadths and moderate ankle edema. Laboratory findings included a venous pressure of 170 mm., negative first and second strength P.P.D., polycythemia (r.b.c. 7.0 million, hgb. 17.5 gm.). On November 13, 1945, pericardectomy was performed by Dr. William P. Longmire, Jr. The left ribs 3 to 5 were resected, and a densely fibrotic process was found involving all aspects of the heart visualized. A large area of both left and right ventricles was decorticated. Following operation venous pressure fell to 75 mm. saline, and fluid accumulation ceased. He was well and doing hard farm work when seen in March and October, 1946. When restudied in December, 1948, the venous pressure was 95 mm. saline; the liver was at the right costal margin. However, on examination of the heart the second pulmonic sound was found to be reduplicated, and there was a mid-diastolic sound at the apex suggesting mitral stenosis to one observer. On auscultation inspiration was conspicuously cog-wheel in character over both sides of the chest but especially on the left. The patient had been actively engaged in farm work when seen in 1951.

Case 17. E. C. P. This white male patient, born in 1895, from July to October, 1945, had a fever of undetermined cause accompanied by night sweats, epigastric fullness and weight loss. Shortly thereafter there was onset of ankle edema, ascites and paroxysmal coughing when

lying on the right side. Physical examination in 1945 revealed marked venous distention, limited chest expansion, particularly on the right, blood pressure 110/85, pleural effusions, ascites, leg edema and liver down four finger-breadths. Venous pressure was 200 mm. saline, circulation time 16 seconds (Decholin). X-ray showed moderate enlargement of the heart, a high right diaphragm, and pleural thickening. On November 7, 1946, pericardectomy was performed by Dr. Alfred Blalock through an incision in the fourth right interspace. The right auricle and ventricle were decorticated. The venous pressure fell to normal in the first post-operative week. On examination in November, 1948, the heart sounds were found to have a peculiar scratchy pericardial quality. Venous pressure was 100 mm. saline. In May, 1950, it was learned that the patient had been working full time as a post office clerk without any difficulty whatever. Heart sounds were perhaps somewhat muffled but otherwise not remarkable. The liver was not enlarged. The etiology in this case was clearly tuberculosis. The scar tissue removed at cardiac decortication contained tubercles. His first wife died of tuberculosis, and in July of 1948 the patient developed tuberculosis of the right ankle. A lymph node from the right inguinal area showed tubercles. In July, 1951, the patient was still well.

Case 18. B. S. This is a white female patient born in 1908 who at the age of 7 had pain in her knees. At the age of 14 she was told she had a "bad heart". From the age of 18 on she required two pillows for sleeping and had fairly marked exertional dyspnea. Her first pregnancy was terminated successfully by Caesarian section at age 27. In 1940 ascites with slight ankle edema appeared, and the patient required 17 or 18 paracenteses in a 3 months period. The significant findings on examination in that year were generalized venous distention, right hydrothorax, absent precordial impulse, reduplicated first sound or presystolic gallop at the apex, liver down 3 finger-breadths, marked ascites, blood pressure 104/80, venous pressure 275 mm. saline. On August 28, 1940, the first operation was performed by Dr. W. F. Rienhoff, Jr. On the left the 3rd and 4th costal cartilages and a portion of the sternum were removed. Numerous adhesions were discovered between the parietal and visceral pericardium and these were easily freed with the finger. A portion of parietal pericardium was resected. Following this procedure the venous pressure remained elevated. Consequently on November 18, 1940, Dr. Rienhoff opened the right side of the chest by an arcuate parasternal incision. The great veins were decorticated. Venous pressure on February 8, 1941, was 165 mm. saline. Throughout a six month hospitalization the patient ran a persistent low grade fever which was not satisfactorily explained. Numerous tuberculin skin tests were negative. A story was elicited of frequent episodes of eczematous eruption over the nose and at one time over the entire body.

When examined in 1943 the patient was able to do the work of a farm wife and sleep on one pillow. The right chest was nearly immovable. The venous pressure was 175 to 200 mm. saline. In the interval until 1949 the patient had felt quite well and had been able to work hard. She had noted ankle edema and dyspnea with increased salt intake. When studied in 1949 significant findings included a twin pregnancy, distended neck veins, pitting edema of the legs, venous pressure 290 mm. saline, vital capacity 1.1 liters in 3 seconds. With digitalization and bed rest the venous pressure returned to normal and the patient was delivered of healthy twin infants at term without complications.

Case 19. P. E. G. This white female patient born in 1916 was in contact with two maternal aunts and a maternal uncle, all of whom died of tuberculosis. At the age of 10 she noted that she had to drop out of games because of dyspnea. Ankle edema progressing to anasarca began at age 12. Digitalis was administered without benefit for about 10 years beginning at the age of 12. In 1941 when the patient was studied at the Peter Bent Brigham Hospital by Dr. C. S. Burwell the principal cardiac findings were a systolic retraction of the left precordial

costal cartilages, a scratching mid-systolic sound, and in the first half of diastole a prominent third sound which at the apex was louder than the first and second sounds. Venous pressure was 215 mm. saline, and total proteins were 3.9 gms. There was pericardial calcification located principally in the atrioventricular sulcus. On November 4, 1941, operation was performed by Dr. Elliot C. Cutler using an arcuate left parasternal incision. Only a partial decortication was possible because of adherence of the pericardial scar to the coronary arteries. Following operation there was moderate but persistent improvement. She has been studied at frequent intervals during the nine years since operation. The loud protodiastolic sound has persisted. Venous pressure has been between 140 and 200 mm. saline. The tendency to accumulate fluid has continued but has been held in abeyance by careful salt restriction and high protein intake. In June, 1950, the neck veins were moderately distended, there was a very prominent early diastolic sound, the liver was 1½ finger-breadths below the right costal margin and there was slight pitting ankle edema. Total serum proteins were 5.6 gms.% with an albumin of 2.5 gms.%. Venous pressure was 170 mm. saline. One pregnancy had been successfully completed in January, 1949, and the patient was again pregnant in June, 1950. This terminated in spontaneous abortion following which the patient felt subjectively improved.

Case 20. E. C. This white male patient, born in 1928, was exposed to tuberculosis as his mother died of this disease when he was 3 years old. At the age of 2½ years he developed tuberculosis of the right hip which required prolonged hospitalization. While the patient was in a body cast at the age of 15 years, dyspnea was first noted. On examination in 1945 findings included blood pressure 107/80, absent apical impulse, split first sound, tic-tac rhythm, marked tachycardia (persistent for about one year), liver down 3 to 5 cm., palpable spleen, ascites and sacral edema. Venous pressure was 260 mm. saline, total proteins 4.81 gms. with 2.25 gms. albumin, P.P.D. positive in the first strength. On April 4, 1945, pericardectomy was performed by Dr. Alfred Blalock through a curved left parasternal incision with removal of costal cartilage 2 to 6. Mainly the right ventricle was decorticated. The pericardium was very adherent particularly along the left anterior descending coronary artery. Histologically the surgical specimen showed only scarring and thickening of the pericardium. By May 2, 1945, serum proteins had returned to normal, but venous pressure was still 230 mm. saline, and the liver was down 3 finger-breadths.

When re-examined in 1950 it was found that the patient had been very well since operation. There was no edema and no dyspnea. His activity had been limited only slightly and that by his orthopedic deformity. Physical examination revealed normal cardiac action as visualized and palpated through the defect in the bony thorax, liver not palpable, no edema, cog-wheel inspirations over the left hemi-thorax only. Venous pressure was 140 mm. saline. On fluoroscopy the heart was normal in size and appearance. There was during inspiration a tugging of the left diaphragm by the heart as a result of an inferoposterior adhesion.

SOME LABORATORY OBSERVATIONS

Cardiac catheterization

Sixteen cardiac catheterizations[1] were performed in thirteen patients in this series. The right ventricular pressure recordings were available for review in eleven catheterizations in ten patients. One of the tracings was recorded with a Hamilton manometer, six with a Statham strain gauge and five with a Sanborn Electromanometer. All but one of the right ventricular pressure tracings showed an early diastolic dip followed by a rapid rise to a diastolic plateau at a high pressure level (as high as 32 mm. Hg in Case 2). All the patients in whom these characteristic pressure curves were recorded either were pre-operative and had a "flat-top and V" pattern by electrokymography (2) or still had clinical evidences of constriction with persistence of the characteristic EKY pattern after operation. In brief, the correlation between the right ventricular pressure pattern and the electrokymographic pattern was close. Unfortunately, however, no pressure tracings are available in patients who had obtained clinical and EKY improvement from operation (2). Hansen and co-workers (3) have described cases in which the diastolic "dip" disappeared following pericardectomy. Case 12 who did not show the characteristic pattern was catheterized before development of the full syndrome of cardiac constriction.

[1] Most of the catheterizations were performed in the laboratory of Dr. Richard J. Bing, to whom the author is indebted for permission to include the data here.

In describing the characteristic right ventricular pressure curve[2] in constrictive pericarditis, both Bloomfield (4) and Hansen (3, 5) emphasized that it is not specific since right sided heart failure of any cause may show an early diastolic "dip" and a diastolic plateau. Richards (6) published such curves in cases in failure and normal curves after compensation. Wood and co-workers (7) published tracings from a case of constrictive pericarditis showing an early diastolic "dip" followed by a diastolic plateau. Two tracings recorded by means of strain gauges were published and these authors interpreted the early diastolic "dip" as a distortion produced by a low frequency recording system since the early diastolic "dip" was less conspicuous (but still present) in recordings by an instrument of higher frequency response. Sawyer (8) has found this pattern quite consistently in constrictive pericarditis but also recorded it in a case of myocardial fibrosis. Hansen (9) likewise found it in one case of myocardial fibrosis and we have recently had the same experience.

Wiggers (10) believes the small early diastolic "dip" in right ventricular pressure curves recorded by cardiac catheterization even in normal individuals is a "low frequency artefact." The inference would appear to be that all such "dips" are artefacts. As stated by Katz (11) in his critical review, the "dip" was described by early workers using cruder recording apparatus and was attributed to a suction action of the relaxing ventricle. It could not be demonstrated in normal animals by more recent workers employing instruments designed according to the principles of Frank.

The presence of a suction action in the normal relaxing ventricle is probably disproven. There is at least a remote possibility that ventricular suction occurs in constrictive pericarditis. Retraction of portions of the chest wall with systole is a conspicuous sign in some patients, e.g., Case 7. Elastic recoil of the rigid

[2] For an example of the characteristic pressure curve, see Figure 9 in an accompanying paper (2).

pericardial case or of the thoracic cage to which it may be adherent might conceivably result in a suction effect.

TABLE 1

Findings of Cardiac Catheterization

NO.	CASE	TYPICAL RT. VENTRICULAR PRESSURE CURVE	PRESSURES (MM. HG.)*				C.I.	ARTERIAL % O_2 SAT.
			RA	RV	PA	PC		
1*	1. (S. W. A.) 10-8-48		28/21	37/20†			3.05	97.4
2*	2. (R. McA.) 1-17-49	Yes (S. G.)‡	37/31	53/32	43/17		2.04	95.0
3*	2. (R. McA.) 5-3-49		10/6	41/8	33/18		3.78	
4	3. (A. N.) 8-30-49	Yes (EMN)		50/20			2.50	90
5*	5. (C. L. M.) 8-10-50	Yes (S. G.)		32/14			2.28	96
6*	6. (L. N.) 12-16-49				42/26		1.27	87
7*	7. (W. B.) 9-7-50	Yes (S. G.)	36/28	57/30			2.06	95
8*	7. (W. B.) 10-9-50		7/3		18/8		2.45	
9	8. (J. M.) 12-29-49	Yes (EMN)	14/7	35/12	38/18	12	2.09	85
10*	9. (N. D.) 5-11-49	Yes (S. G.)	17/11	31/13	24/13		1.9	98
11*	9. (N. D.) 11-22-49	Yes (S. G.)	19/10	30/6	30/13	10	3.91	91
12*	10. (W. M.) 5-25-50	Yes (S. G.)	18/12		42/18	24	2.73	87
13*	12. (M. T.) 9-17-48	No (S. G.)	30/26	31/16	31/16		2.0	
14	13. (J. H.) 4-27-50	Yes (EMN)	28/17 (mean 24)	50/15 (mean 35)	45/28	28	2.0	100
15*	14. (G. V. S.) 6-4-47	Yes (Ham.)	33/28	49/29	44/31		2.10	96
16*	15. (P. H.) 4-9-48						2.14	96

* Study performed in laboratory of Dr. Richard J. Bing.

† RA = right auricle. RV = right ventricle. PA = pulmonary artery. PC = pulmonary "capillary". (In reporting ventricular diastolic pressure the level of the diastolic plateau was used rather than the pressure at the nadir of the early diastolic "dip".)

‡ S. G. = Statham strain gauge. Ham. = Hamilton manometer. EMN = Sanborn Electromanometer.

The mechanism of the right ventricular pressure pattern is not entirely clear. However, until further evidence to the contrary is available, the writer prefers to consider the "characteristic" right ventricular pressure curve as a reflection

of an actual cardiodynamic event. The early diastolic "dip" corresponds closely with anomalous cardiodynamic phenomena recorded by sound tracings (2, 16) electrokymograms (2), and ballistocardiograms (12). That the pressure pattern may be bona fide is further supported by the finding of Dr. Hansen (3) while working at the Johns Hopkins Hospital. In Case 7 early diastolic "dips" were recorded from both left and right ventricles through needles introduced directly into the ventricle at operation. Furthermore, the early diastolic dip was of roughly the same absolute amplitude in the two ventricles.

In constrictive pericarditis the right auricular pressure pulse is wider than it is normally, that is, the auricular pulse pressure is greater. Furthermore, at the time of ventricular filling there is a deep V-shaped excursion of the auricular pressure curve. The complete curve is roughly M-shaped. The wide pulse pressure is probably explicable on the basis of the electrokymographic findings (2). With auricular contraction pressure rises higher than usual since ventricular filling is already complete. With rapid ventricular filling the auricular pressure drops relatively low for a brief period. The right auricular pressure curve of constrictive pericarditis provides two bits of evidence suggesting that the "characteristic" ventricular pressure curve is bona fide (cf. (2) for tracings):

(A). A "dip" occurs in the right auricular pressure curve at the same time as in the right ventricular pressure curve;

(B). The auricular and ventricular "dips" are of roughly identical shape and absolute amplitude.

The elevated "pulmonary capillary" pressure in those instances where it was determined indicates the importance of the factor of left ventricular constriction in these cases. This finding is in accord with those of Burwell (37).

Angiocardiography

Angiocardiography was performed in four of these cases (# 1, 3, 8, and 15). In none was a localized inflow obstruction demonstrated. Sellors (41) expresses the opinion that the venae cavae "can be reduced to the diameter of little more than a lead pencil without interfering with an efficient circulation, providing that the ventricles are sound." The positive findings of angiocardiography included: (1) dilated superior vena cava (all cases); (2) thickening of the wall of the heart on the right border (right auricle) in Cases 1 and 15; (3) close apposition of the opacified right ventricle to the sternum as seen in lateral view (i.e. obliteration of the retrosternal space) in Cases 1, 8, and 15; (4) dilated main pulmonary artery in Case 8; and (5) dilated left auricle in Case 8.

Renal clearances

In seven of these patients determinations were made of glomerular filtration by inulin or thiosulfate clearance, of effective renal plasma flow by para-aminohippurate clearance, and of filtration fraction by derivation. The results

are given in Table 2. The most conspicuous feature is a consistent elevation of the filtration fraction. Further elevation of this value with exercise was observed in the two cases in which this was tested. Elevation of the filtration fraction occurs with congestive heart failure due to other causes (13).

TABLE 2

*Renal Clearance Data**

CASE	DATE	GLOMERULAR FILTRATION RATE	RENAL PLASMA FLOW	FILTRATION FRACTION
1. (S. W. A.)†	10–6–48	127	610	.21
		168	832	.20
		103 } exercise on treadmill	390	.26
		126 } exercise on treadmill	445	.28
		133	520	.26
		150	557	.27
		124	458	.27
3. (A. N.)	9–5–49	104	318	.33
		82	259	.27
9. (N. D.)	5–13–49	145	473	.29
		136	259	.27
12. (M. T.)†	9–9–48	73	168	.44
		73	170	.46
		76	167	.47
		78	135	.58
		69	215	.32
13. (J. H.)	6–7–50	125	468	.27
		107	344	.31
14. (G. V. S.)†	5–29–47	110	364	.30
15. (P. H.)†	4–19–48	82	310	.27
		76	260	.29
		87	264	.33
		78 } exercise on treadmill	266	.29
		61 } exercise on treadmill	194	.32
		80	232	.35
		83	242	.32
		73	249	.34

* All 12 to 16 minute periods. Consecutive periods when more than one given.

† Data provided by Dr. Elliot V. Newman.

GENERAL CLINICAL COMMENTS

Etiology

In eight of these twenty cases there was adequate evidence for a tuberculous etiology. The evidence consisted of unusually concentrated and prolonged exposure to tuberculosis or evidence of tuberculosis in the pericardial scar or at

sites other than the pericardium, e.g., bone, lymph node, or lung. The other twelve cases are representative of the majority of cases, in which evidence of a specific etiology is unobtainable. This does not necessarily indicate that tuberculosis is not the cause in most if not all cases. Even the absence of more than non-specific scar at operation or autopsy is no evidence against tuberculosis. A very instructive case in this regard was studied at the Johns Hopkins Hospital over a 20 year period and has been reported elsewhere (19). At various times during this period of observation he had clinically typical tuberculous pericarditis with effusion, cervical lymphadenopathy which was proved to be tuberculosis by biopsy, active pulmonary and mediastinal tuberculosis, pleural effusion with acid-fast bacilli demonstrable in the fluid, and presumably tuberculous chorioretinitis. In 1935 he developed the full-blown syndrome of chronic constrictive pericarditis with calcification by x-ray. In Cleveland on July 25, 1936, Dr. Claude S. Beck performed cardiac decortication; in spite of the unequivocal past history, only nonspecific hyalinized and calcified pericardial scar was found. Case 20 is very comparable. There was undoubted tuberculosis of the hip, but the pericardial lesion was only non-specific scar at the time of operation.

There is a significant number of cases with typical constrictive pericarditis in whom the tuberculin skin tests are repeatedly negative. Cases 14, 16, and 18 in this series and patient C. W. described later are examples of this. Speculation as to the etiology of these cases I will leave to others. It has not been possible to discover in the pathology files of the Johns Hopkins Hospital a case of constrictive pericarditis conceivably due to disseminated lupus erythematosus (42).

Trauma should not be overlooked in the etiology although this cause has never been proved unequivocally. Blunt trauma to the chest, such as the "steering wheel injury" of Beck (20), can produce hemopericardium with subsequent organization and calcification and, conceivably constriction. It is very likely that Case 5 was due to trauma because of the finding of a subdural hygroma-like cyst between the heart and the sternum and because of the history of profound although indirect trauma two years previously. By analogy to subdural hygroma the abrupt and relatively late onset of symptoms is readily explicable. The subjective complaint of "fullness of the head" in the supine position was quite out of proportion to that usually observed and may be explained by the nature and position of the pericardial cyst in this case. Mortensen and Warburg (17) report a case of this condition due to trauma. It is possible that trauma to the chest may precipitate constrictive pericarditis by causing rupture of tuberculous lymph nodes into the pericardium with subsequent scarring. White and Glendy (21) described what may be such a case: "In one of our 16 cases of Pick's disease a severe blow on the chest preceded the symptoms and signs of a bloody pericardial effusion, which later developed into a chronic tuberculous constrictive pericarditis requiring operation."

The protodiastolic sound

An adventitious sound in early diastole was described at some time in about half the cases of constrictive pericarditis in this series (Fig. 1). Simultaneous recordings of heart sounds and ventricular border electrokymograms conclusively demonstrate that the protodiastolic sound is due to rapid ventricular filling early in diastole and/or abrupt halt in ventricular filling (2).

The protodiastolic sound of constrictive pericarditis was apparently first described by Potain (14) in 1856. Lian, Marchal, and Pautrat (15) wrote about

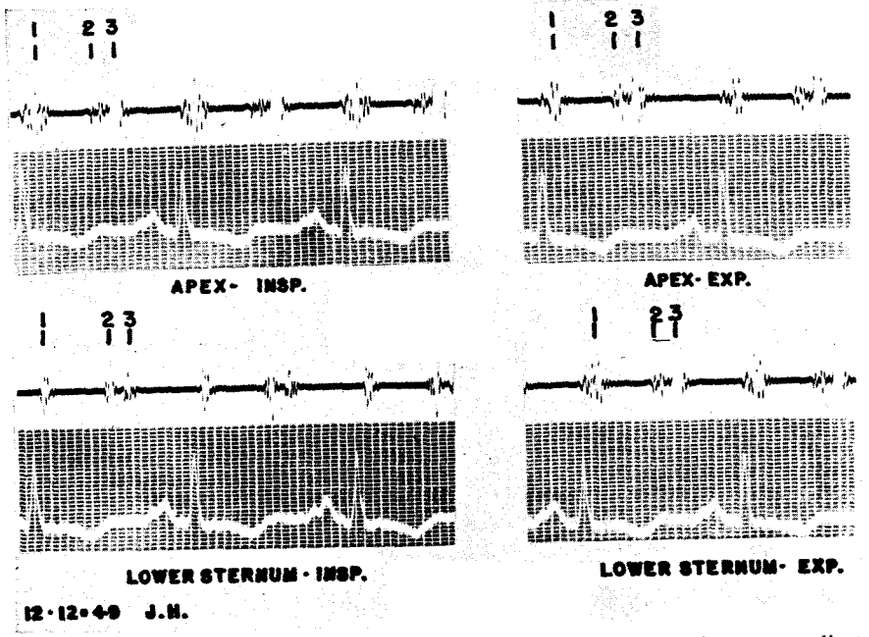

FIG. 1. These phonocardiograms from patient #13 show the prominent protodiastolic sound (marked "3"). At the apex during inspiration and at the lower sternum during expiration the protodiastolic sound is louder than the first and second sounds.

this protodiastolic sound and referred to it as the pericardial protodiastolic vibration to distinguish it from the normal third heart sound and protodiastolic gallops. (This terminology has advantages since the term "gallop" has well established diagnostic and prognostic connotations.) They believe that it occurs only with pericardial calcification. This indeed appeared to be almost true in our studies as well. Only Case 6 had a definite protodiastolic sound in the absence of pericardial calcification. Why calcification should be necessary for the development of the extra sound is not clear since calcification is not essential to its explanation (2).

Mortensen and Warburg (17) found a "mesodiastolic gallop" in 16 of 25

cases. Paul, Castleman, and White (18) observed "a distinct third sound" in 15 of 53 cases.

Serum proteins

Total serum proteins and albumin-globulin ratio were determined at least once in all twenty cases of constrictive pericarditis. Cases 6, 16, 19 and 20 had serum albumins of 3.0 gms. or less. Cases 13 and 14 had serum albumins between 3.0 and 3.5. All six of these cases had had their disease from at least the early teens. Pediatricians in particular (22, 23) have emphasized the serum

FIG. 2. The "sugar-coated" liver in a dog with chronic constrictive pericarditis produced by cellophane around the heart.

protein changes in the syndrome of chronic constrictive pericarditis. Growth may place demands on the congested liver which it cannot fulfill but which are not present in the adult. Another possible factor may lie in the fact that few of these patients had any abdominal paracenteses. In the patients who were tapped, paracenteses were performed only two or three times at widely spaced intervals. Regeneration of serum proteins may occur very rapidly after operation as seen in Case 6.

The liver

Five patients in this series had hepatic friction rubs synchronous with respiration. Palpation of the enlarged liver in one of these cases produced a pecu-

liar sensation to the examining hand somewhat resembling subcutaneous emphysema. This is the clinical analogue of the *Zuckergussleber* of Curschmann (24). A dog[3] in which the clinical syndrome of constrictive pericarditis was produced by wrapping the heart in cellophane showed at autopsy six months later marked "sugar coating" of a much enlarged liver (Fig. 2). Histologically this coating was fibrin overlying a collagenous thickening of the liver capsule (Fig. 3). Blalock and Burwell (25) described "hyperplastic peritonitis" in their dogs with aleuronat pericarditis. In patients the hepatic friction rub tends to occur post-operatively while the liver is shrinking in size and may be a source of much discomfort to the patient and concern to the physician. It appears, however,

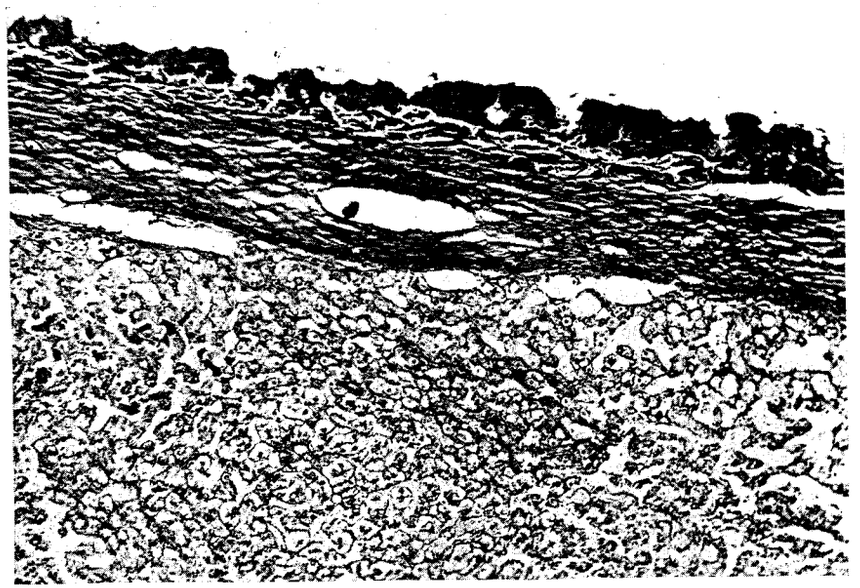

FIG. 3. Histological appearance of "sugar-coated" liver shown in Figure 2. There is fibrous thickening of Glisson's capsule with much overlying fibrin.

to be of no serious import and most of the patients who have shown it have had clinical cures.

Chest x-rays

The dilated superior vena cava is an impressive feature of the chest x-rays of these cases. Before operation it was present in some degree in all twenty cases of the constrictive syndrome. Venous pulse type electrokymograms (2) are usually easily recorded from the dilated structure. In few other cardiac conditions does elevated venous pressure persist for such a long period of time.

[3] Reported through the courtesy of Dr. James O. Davis of the National Heart Institute.

As a result the elasticity of the vena cava is lost just as is the case with the neck veins. A dilated superior vena cava with little or no enlargement of the heart is highly suggestive of constrictive pericarditis. Among others, Kerley (26) and Friedberg (27) have remarked on this phenomenon.

Calcification was demonstrated roentgenographically in 10 of the 20 cases of constrictive pericarditis in this series. This is approximately the percentage of calcification found in the series of 53 cases published by Paul, Castleman, and White (18). The calcification was always predominant on, or limited to, the diaphragmatic aspect of the heart. Presumably this is due to sedimentation of inflammatory elements which later become calcified. The calcification also tends to be dense in the atrio-ventricular grooves. The apex is likely to be free of calcification.

Respiratory function

Impaired respiratory function of a type pointing to primary pleuro-pulmonary disease is present in most cases, outstanding in some. They usually have thickened pleura by x-ray, reduced vital capacity, impaired chest expansion. Most cases, one must conclude, have some degree of constrictive pleuritis in addition to constrictive pericarditis. An extreme example of this association was reported by Burwell and Ayers (28). Recently at the Johns Hopkins Hospital a case of constrictive pericarditis with calcification of the pleura as well as the pericardium has been studied.

Cyanosis

Cyanosis with arterial oxygen unsaturation at rest is uncommon but not unknown in constrictive pericarditis (29, 30, 31). Mortensen and Warburg (17) reported cyanosis in 18 out of 25 cases; there were no data on arterial oxygen saturation. Cases 7, 9, 12, 13, and 16 were cyanotic at rest and Case 14 became cyanotic with even mild upper respiratory infections. (In Case 10 the cyanosis was probably due to methemoglobin which was found to represent 10% of the total hemoglobin and which may have been produced by mannitol hexanitrate.) Cases 7 and 16 had polycythemia as well as cyanosis. Friedberg (31) suggests that the probable cause is "associated pleuropulmonary disease or peripheral venous stasis."

Syncope

Syncopal attacks occurred on at least one occasion in Cases 2, 10, and 16. In all three cases the syncopal attacks occurred during exertion. Since these cases are prone to auricular arrhythmias, a bout of tachycardia or cardiac standstill preceding change in mechanism may be the cause. However, in Cases 10 and 16 fibrillation or other arrhythmia have never been observed. Failure of the cardiac output to increase adequately with exercise (31) is probably a more likely

explanation for the syncope. In Case 2 syncope occurred during electrokymography (2). When the patients hold their breath, as is required in this procedure, they frequently perform a Valsalva maneuver. Syncope occurs in cases of superior vena cava obstruction when venous return through what channels remain is reduced by some factor such as the Valsalva maneuver (32). These observations suggest that straining may be a factor in the syncope of constrictive pericarditis.

Simulation of mitral stenosis

Physiologically and sometimes roentgenologically constrictive pericarditis can very closely simulate mitral stenosis. It is clear that from the physiologic standpoint it makes no difference whether the left ventricular filling is impeded by narrowing of the mitral orifice or by a decrease in diastolic capacity of the ventricle itself. In both conditions a high pulmonary "capillary" pressure is recorded by catheterization. Case 8 (J. M.) had in addition to an elevated pulmonary "capillary" pressure a mammoth left auricle presenting far over on the right side of the heart. Others of these cases, e.g., Cases 2 and 7, had lesser degrees of left auricular enlargement. Not included in this series are two Johns Hopkins Hospital cases of constrictive pericarditis who demonstrated left auricular enlargement to a most pronounced and confusing degree. C. W., a 22 year old white male patient, had the typical history and physical and x-ray findings of constrictive calcific pericarditis. In addition there was a large dog-ear-like mass projecting from the mid portion of the left border of the cardiovascular silhouette. Exploration which was performed because of suspicions of tumor disclosed a greatly dilated left auricular appendage. R. S., a 19 year old colored male patient who was likewise a classic case of constrictive pericarditis, showed radiographically great enlargement of the left auricle to the right and posteriorly displacing the esophagus and left main bronchus. The main pulmonary artery was very prominent due apparently both to dilation and to displacement by the large left auricle. These findings were confirmed by angiocardiography and by surgical exploration at the time of pericardectomy.

Enlargement of the left auricle is clearly demonstrated in x-rays published by White (33). Involvement of the left side of the heart in constrictive pericarditis has been emphasized by White, Alexander, Churchill, and Sweet (34), and by Sawyer (8). Rarely the characteristic auscultatory and cardiodynamics changes of mitral stenosis may be produced by penetration of the atrio-ventricular fissure by the fibrosing and calcifying process with involvement of the mitral ring (16, 38).

One physiologic distinction between mitral stenosis and constrictive pericarditis lies in the response of cardiac output to acceleration of the cardiac rate. In mitral stenosis cardiac output bears, within limits, an inverse relationship to cardiac rate; in constrictive pericarditis the relationship is usually a direct one (30).

Constrictive pericarditis may simulate tricuspid stenosis by virtue of the presence of a presystolic or atrial liver pulsation as pointed out by Wenckebach (35). The fact that the capacity of the ventricle has been reached when the auricle contracts is presumably the mechanism (2).

Results of pericardectomy

Of the twenty cases of constrictive pericarditis in this series seventeen have been subjected to at least one pericardectomy. Fifteen of the cases have been observed for at least two months following operation. The results of operation are given in Table 3. Based on this experience it is the opinion of the writer that every case of constrictive pericarditis with the probable exception mentioned below should be subjected to pericardectomy as soon as the diagnosis is established, active tuberculosis excluded or at least controlled with chemotherapy, and the patient in as good a physical condition as he can be expected to attain. All this assumes, of course, that the patient's symptoms are incapacitation.

TABLE 3

Results of Pericardectomy

POST-OPERATIVE CONDITION	TOTAL NUMBER OF CASES	NUMBER OF INDIVIDUAL CASE IN THIS SERIES
Dead	2	8, 10
Improved	3	3, 13*, 14*, 19
Clinically cured	11	1, 2, 4, 5, 6, 7, 15, 16, 17, 18, 20

* These may be clinical cures.

Two deaths in this series occurred while the patient was on the table undergoing pericardectomy. One patient in this group died without operation. These were the three oldest patients in the series (with the exception of Case 5 which was probably traumatic in origin). In addition these three patients had *Panzerherz*, i.e., very extensive pericardial calcification. Operation should probably not be attempted in individuals with the combination of relatively advanced years and extensive calcification (36). There is likely to be so much myocardial atrophy or coronary artery disease that rupture of the heart or cardiac standstill will occur as was the case in our two patients. By way of contrast cases such as Cases 1 and 5, although in the older age group at the time of onset of symptoms, did not have pericardial calcification and obtained clinical cures from pericardectomy. In the younger age group good results can be expected as a rule in spite of pericardial calcification.

One patient (Case 13) appears to have attained a clinical cure one year after the last of three cardiac decortications. Case 14 has likewise had three operations and although she is now much improved it is still too early to be certain about the ultimate result. What is the cause of the delayed or incomplete success in these fortunately infrequent cases? It seems likely that these cases

have fibrosis involving the myocardium in a too intimate manner to permit complete removal by the surgeon. Isaacs (40) has experimental data in support of this idea.

It will be noted from perusal of the individual case reports that there has been an evolution[4] of incisions or surgical approaches to the pericardium as indicated by Dr. Blalock in the introduction to this group of papers.

SUMMARY AND CONCLUSIONS

1. Clinical features of twenty cases of constrictive pericarditis are presented.

2. A case is presented (Case 5) in which trauma may have been the main etiologic factor in the scarring of the pericardium.

3. Cyanosis, impaired respiratory function, syncope, dilatation of the superior vena cava, serum proteins, simulation of mitral stenosis, and the hepatic friction rub are discussed.

4. Angiocardiography was of no significant diagnostic aid in four cases.

5. Elevation of the filtration fraction was a consistent feature of renal hemodynamics in seven cases studied by renal clearance techniques.

6. In thirteen patients cardiac catheterization was performed. An early diastolic "dip" followed by a diastolic plateau was a consistent feature of the right ventricular pressure curve. All evidence available from these studies is consistent with the view that these recordings represent a bona fide cardiodynamic phenomenon.

7. An adventitious sound in early diastole was present in at least half of the cases in this series.

8. Results of pericardectomy were in general satisfactory. Of fifteen cases who survived operation, eleven attained clinical cures, three were improved, and one unimproved. Two patients died during operation from ventricular rupture and cardiac standstill, respectively. Both patients were over 55 years of age and had very extensive pericardial calcification.

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[4] On the surgical service of Dr. Alfred Blalock where most of these cases were operated upon.

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